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NOTES ON SOME CASES OF CONGENITAL MALFORMATIONS OF THE FACE AND EARS.

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Case I.—A. H., aged 23 years, whom I had the privilege of examining by the courtesy of Dr. H. S. Straight, presents several interesting congenital defects. There is no deformity in either of his parents, nor in any of his brothers or sisters.

On the right cheek there is a circular depression one and one-half inches behind the angle of the mouth. This depression or dimple is one-eighth of an inch in depth and one-sixth of an inch in diameter. One and one-half inches above it, just in front of the tragus, there is another depression, rather larger than the former.

The right external auditory meatus is entirely absent. Corresponding to the position of the tragus there is an irregular mass which projects three-fourths of an inch from the surface, and is about an inch long. In its interior it evidently contains cartilage. The auricle is otherwise well formed.

The right ramus and angle of the mandible are smaller than the corresponding parts on the left side. All the teeth in the inferior dental arch are present. The temporo-mandibular articulation and the coronoid process are apparently normal.

On the left cheek there is also a fistulous opening leading to a canal, about one inch long, which passes subcutaneously towards the auricle. According to the patient's statement, this opening was at one time larger than at present, and from it issued a sticky fluid. The left pinna is small and deformed. There is an opening, leading to a short blind canal, at the extremity of the helix, and a supernumerary auricle exists in front of the tragus. The accompanying illustrations well represent the appearance of the patient. In a subjoined note, by Dr. Straight, the aural and pharyngeal condition are further described.

Upon examining the patient for other peculiarities, we find the breasts to be rather large and the lower portion of the abdomen rather protuberant, giving rise to an appearance approaching that of a female.

Near the end of the coccyx is a somewhat funnel-shaped depression, into which the tip of the little finger can be placed.

Case II.—Mrs. A., whom I saw by the courtesy of Dr. Dudley P. Allen, has a deformed auricle on the right side. Near the top of the pinna there is a shallow circular depression. The meatus auditorius externus is entirely absent. She fails to hear the ticking of a watch when applied to the side of the head. On the left side there is a small supernumerary tragus. No other defects exist. Her father presents almost identically the same deformity of the right ear.

To explain the facial deformities in the first case we must assume some grave defect in the formative processes in connection with the first visceral arch and cleft. The dimples and fistulous tract in the cheek may be accounted for, according to Bland Sutton, and others, by assuming an irregularity in the closure of the mandibular fissure. This fissure, which is between the superior and inferior maxillary processes, is relatively much wider in early embryonic life. It closes, however, in its posterior or dorsal portion, leaving the anterior portion open, and thus forming the mouth. When the fissure fails to close to the proper extent, the condition known as macrostoma results. When, on the contrary, the margins unite too extensively, microstoma results. Irregularity in the closure may, as above stated, lead to the production of dimples and fistulous tracts in the cheek.

As His[1] has shown, the pinna is formed by the fusion of six tubercles, which surround the opening of the first branchial cleft or furrow. Defective fusion of two or more adjacent tubercles may, as Sutton suggests, result in the production of an auricular fistula. An irregular and aberrant development of the tubercle forming the tragus may account for the fibro-cartilaginous mass occupying the position of the tragus.

The external opening of an auricular fistula may become occluded, and subsequently, from the accumulation of the secretion of the epithelial lining of the canal, a cyst may form. Lannelongue[2] pictures such a case, and I have seen a similar one during the past winter. That rudimentary forms of the pinna are sometimes associated with failure in the proper development of one or both sides of the lower jaw is a fact first alluded to by Allen Thomson.

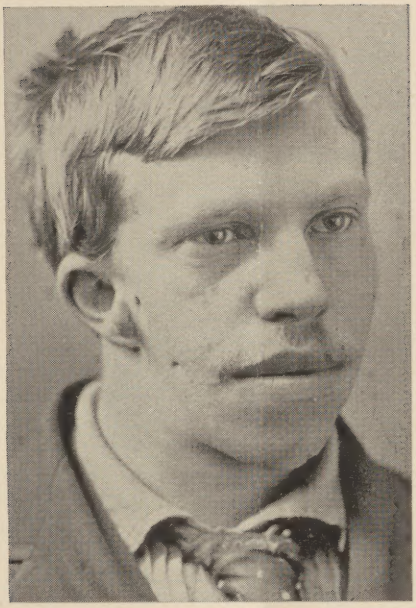
In connection with the depression near the tip of the coccyx, a few remarks may not be out of place.

Ecker,[3] in an elaborate and interesting article, has described this depression (though it had previously been referred to by Luschka), and gives his view as to its origin. This dimple, or *foveola coccygea*, as he terms it, is seen not infrequently in infants and occasionally in adults.

When the skin over the sacral and coccygeal region is removed

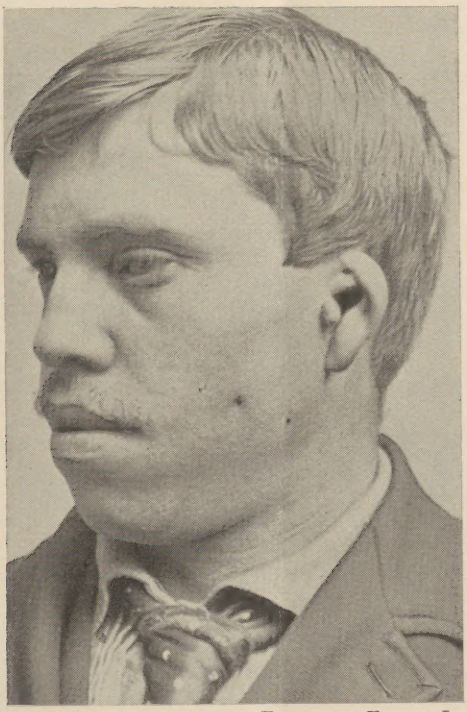
[1] Anat. Mensch. Embryonen.
[2] Affections Congénitales, par Lannelongue et Menard, Paris, 1891, t. I, p. 221.
[3] Archiv f. Anthropologie, Bd. XII.

FIG. 1.



CONGENITAL MALFORMATIONS OF FACE AND EARS. Right side.

FIG. 2.



CONGENITAL MALFORMATIONS OF FACE AND EARS. Left side.

carefully from the subjacent parts, there will be found extending from the tip of the coccyx to the overlying integument a band of fibrous tissue, which was described by Luschka,[1] under the name of the *ligamentum caudale* or *ligamentum apicis coccygis*. Ecker states that in cases in which the foveola is conspicuous, this "ligament" is attached to the bottom of the depression.

In human embryos of four to six months there is a distinct tail-like projection of the posterior or lower end of the vertebral column, as the researches of His, Keibel, and others have shown. Subsequently, with the increased curvature of the sacrum and other changes, this projection disappears; but even in later fetal life, and indeed in young children, the axis of the pelvis is less curved than in adults; in other words, the curvature of the sacrum and coccyx is less marked.

From the fact that there is a connection between the tip of the coccyx and the overlying integument through the medium of the ligamentum caudale, it may be reasoned that the increase in the curvature of these bones has caused a drawing-in of the integument over the tip of the coccyx, producing the so-called foveola coccygea.

A case reported by M. Braun, quoted by Wiedersheim,[2] in which there was a distinctly projecting tail, as well as a foveola coccygea, militates somewhat against Ecker's view. Another explanation that has been given for the mode of origin of the foveola is that there is an imperfect fusion of the two sides of the body, resulting in the production of the depression. It is stated that the depression might extend more deeply, becoming separated from the skin; a closed sac might thus be formed, and from the proliferation of epithelial tissue in its interior a dermoid cyst result.

Further Note Concerning Case I.

A. H. presented himself at the dispensary, complaining of his throat. He had a hypertrophic rhinitis, also a deviation of the septum that nearly occluded the left nasal passage, and a marked inflammation of the naso-pharynx, pharynx, and larynx. The deviation of the septum was removed with a Bosworth saw. The hypertrophies were cauterized with the galvano-cautery and astringent applications made to the pharynx and larynx. The condition of his ears was interesting. In cases like this, in which the auricles are deformed, the rule is that the external auditory canal and the middle and internal ear show most marked changes from the normal on the side of greatest auricular deformity. This was not true in his case. The left ear, aside from the changes in the auricle already mentioned, was normal, except that

[1] Anat. des Beckens.

[2] Der Bau des Menschen, etc., Freiburg, 1893, pp. 24.

the external auditory canal was narrower than ordinarily. This narrowing was due to an exaggeration of the normal curve in the floor of the bony portion. He had an inflammation of the left middle ear, which rapidly improved under the ordinary treatment. Upon the right side he had no external auditory canal. The position of the canal was occupied by bone, in which no sinus or depression could be detected. He could hear a watch upon aural contact only. The watch was heard more distinctly upon mastoid contact. He could not hear a tuning-fork through the air. The bone conduction, however, seemed to be perfect. Upon using Politzer's air-bag, he said he could feel pressure in the ear. Upon using the Eustachian catheter, he said he could feel the air deep in his ear, and that it felt cold. Inflation with air-bag or catheter made no difference in his hearing. He had had a tinnitus, however, at times for two years, and this condition was relieved by the treatment. He probably has upon the right side a complete development of the ear with the exception of the external auditory canal. It seems reasonable, at least, to infer that he has an internal and a middle ear. What is the condition of the tympanic membrane and the auditory ossicles is a question.

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